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OMPHALOCELE : A RARE CASE REPORT

KEYWORDS

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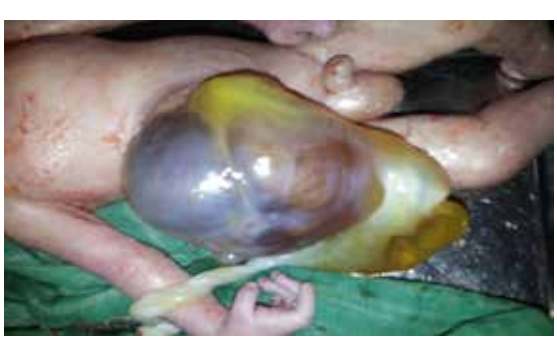
ABSTRACT *An omphalocele is a ventral defect of the umbilical ring resulting in herniation of the abdominal viscera. Omphaloceles occur in 1 in 3,000 to 10,000 live births. Associated malformations such as chromosomal, cardiac, or genitourinary abnormalities are common.*

Postnatal management includes protection of the herniated viscera, maintenance of fluids and electrolytes, prevention of hypothermia, gastric decompression, prevention of sepsis, and maintenance of cardiorespiratory stability followed by primary or staged closure of defect. Here we report a case of large omphalocele containing liver, spleen in addition to bowel.

Case report

A 28 year old woman, gravid 2, para 1 presented to our hospital at 38 weeks period of gestation. She had labour pains since 5 Hours . She was an unbooked case but took iron and folic acid supplementation with 2 doses of tetanus toxoide by anganwari worker.She did not had any antenatal ultrasonography or fetal scan till date. On per abdomen examination she had findings favouring of polyhydramnios and fetal parts were difficult to appretiate. On per vaginal examination cervix was 8 cms dilated, 80-90% effaced with well formed bag of membrane , vertex at -1 station and pelvis was adequate. The patient gave birth to a male child weighing 2.5 kg by vaginal delivery. The child had large omphalocele containing intestines, liver and spleen. These all organs were enclosed in single sac. The child died 10 minutes after delivery.



Discussion

Omphalocele and gastroschisis are the two most common major congenital abdominal wall defects.1Omphalocele is a midline anterior abdominal wall defect with extrusion of abdominal viscera, covered by a membranous sac, into the base of the umbilical cord.Contents of omphalocele are bowel, liver, or both of these. Spleen is rarely a content of omphalocele.2,3

The incidence of omphalocele ranges between 1.5 and 3 per10,000 births.4-6 Patients with omphalocele have a very high (up to 50%–70%) incidence of associated anomalies and high incidence of mortality.7-10 The anamolies of cardio-vascular, genito-urinary, central nervous system and skeletal system are commonly seen with omphalocele.11The incidence of associated anomalies is lower in live born patients because those who have multiple and serious anomalies are more likely to be stillborn.12Chromosome anomalies, notably trisomy 13, 14, 15, 18, and 21, are present in up to 30% of cases. Cardiac defects are also common, being present in 30% to 50% of cases.In many studies mortality was associated considerably with severity of malformations whereas association with chromosomal anamolies was statistically not significant.

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Some investigators have found that size or the contents of omphalocele are of important prognostic value.8,9

The exact mechanism leading to omphalocele is not clear. It has been suggested that, during the 6th week of embryonic development, the rapid growth of bowel causes the intestinal loops to enter the extraembryoniccelome. These bowel loops return to embryonic cavity by 3rd month of gestation. If due to some reason this returning process fails to occur, then this results in omphalocele formation.7At birth contents of omphalocele are covered with amnion, parietal peritoneum and a thin layer of connective tissue.

Omphalocele should be differentiated from Gastroschisis, which is a small defect in the anterior abdominal wall typically located to the right of the umbilical ring and resulting in the herniation of the abdominal contents, without a surrounding membrane, into the amniotic cavity. In gastroschisis, the incidence of associated anomalies is between 10% and 20%, and most of the significant anomalies are in the gastrointestinal tract.5 About 10% of babies with gastroschisis have intestinal stenosis or atresia that results from vascular insufficiency to the bowel at the time of gastroschisis development or, more commonly, from later volvulus or compression of the mesenteric vascular pedicle by a narrowing abdominal wall ring.6 chromosomal abnormalities, are unusual. Gastroschisis is thought to result from an ischemic insult to the developing body wall.12

Prenatal ultrasound could potentially identify the overwhelming majority of abdominal wall defects and accurately distinguish omphalocele from gastroschesis. This identification would permit an opportunity to counsel the family and to prepare for optimal postnatal care.12

The mode of delivery for a known omphalocele containing fetus is a controversial topic. How et al have reported that these foetuses can be safely delivered by vaginal route13 but other studies show improved outcome for fetus with abdominal wall defect delivered by elective caesarean section.14

Still the outcome of patients who have omphalocele depends largely on the associated anomalies and medical conditions and large omphaloceles containing spleen and liver with intestine and other anamolies have higher chances of poor fetal or neonatal outcome. Pregnancy termination options should be discussed with the family depending upon the gestational age of the fetus, and if the family elects for continuation of pregnancy , serial ultrasonography for fetal growth monitoring and any evidence of fetal compromise should be done. In this condition elective caesarean section in presence of neonatal expert and with facilities for immediate pediatric surgical management should be done.

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